Enrico M. Chiappa
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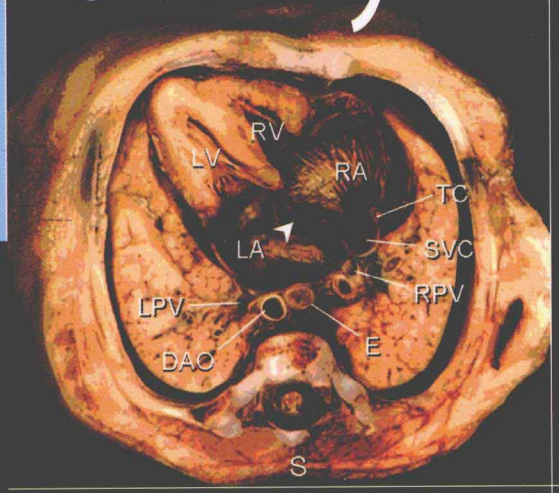
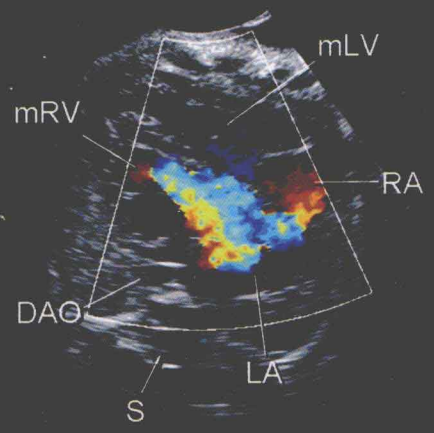
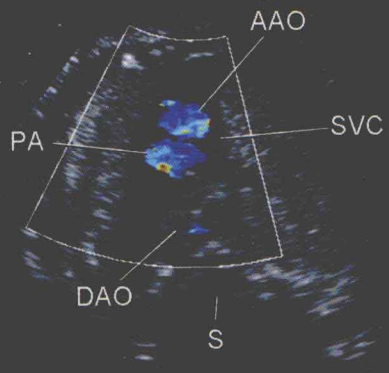
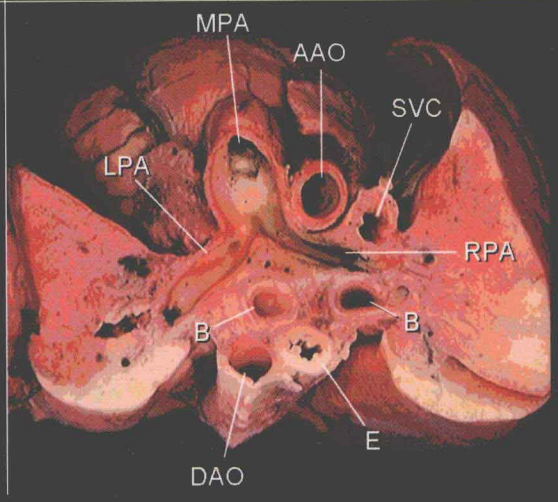
Echocardiographic Anatomy in the Fetus









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To our parents, who paved the way of our lives

Preface

by Norman H. Silverman

Over the last two decades, the value of examining the fetal heart has moved from an experimental procedure of diagnostic curiosity to a front-line form of evaluating fetal cardiac health and disease. There have been numerous advances in the associated technology, including high-resolution imaging, the introduction of reliable color flow and pulse Doppler, and M-Mode and continuous-wave Doppler recordings in some instruments. Such advances continue, with the potential for 3D imaging using spatiotemporal image correlation (STIC) and full-volume fetal technology.

The techniques used by obstetric sonographers in all fields, including physicians from the fields of radiology, obstetrics, and pediatric cardiology, together with technologists who support and do most of the scanning, require a fundamental understanding of ultrasound as well as anatomy, physiology, and the various cardiac pathologies that occur in the fetus. This book addresses these fundamentals, providing correlations by means of diagrams and images of fetal cardiac morphology and pathology. The scans are quite unique, having been collected over several years by the principal author, Dr. Enrico M. Chiappa, from his laboratories in Italy, and provide exquisite echocardiography of normal and congenitally malformed hearts. These are complemented by the excellent pathological images of Dr. Andrew C. Cook and Dr. Gianni Botta, who provided high-quality images of normal and pathological fetal heart conditions, which are displayed as support for the echocardiographic images.

The organization of this book is oriented toward practitioners. The first section provides general guidelines for imaging the fetal body and heart, for segmental analysis, and for diagnosis. The second section takes a view-oriented approach, describing first the transverse views and then the longitudinal views of the fetal body and how each echocardiographic projection best displays a particular pathological entity. The third section contains essential information pertaining to the new technique of 3D/4D echocardiography and the role of the pathologist in heart disease, which expands further the value of this text for providing references and comparisons with standard imaging techniques.

The authors obviously gave a great deal of thought to this project – from the choice of images in the text, which include the clearest descriptions and labels, to the accompanying DVD, which contains complex moving echocardiographic images. The objective is to provide the reader with something greater than a static representation of the fetal cardiac morphology while retaining the ability to refer directly to morphological comparisons and consult with the text for greater detail. This work will have great appeal to physicians and technologists involved in obtaining and interpreting such images and will provide the obstetric, cardiological, and radiological communities with an excellent reference for comparing cases seen in their daily practice.

Preface

by Enrico M. Chiappa

The number of congenital heart diseases detected in utero is still low, even in countries with well-advanced screening programs. Only 20% of new cases of congenital heart disease come from traditional high-risk pregnancies. Ultrasound screening of all pregnancies is therefore necessary to improve the detection rate of congenital heart diseases and to manage them most effectively. However, prenatal screening studies have shown widely divergent results, with low detection rates in most cases. Several reasons have been advocated to explain why these programs seem to fail, and different solutions have been proposed to improve the success rate. There is a broad consensus that examiner skill plays a crucial role in this setting and that appropriate training is mandatory.

It is our belief that people responsible for fetal cardiology units in centers of excellence should invest resources to fully educate the personnel involved in prenatal ultrasound screening. To do this, a fundamental step is to improve, simply and comprehensibly, their knowledge about the anatomy of the fetal cardiovascular system. As Prof. Robert Anderson beautifully stated in a recent review [Anderson RH, Razavi R, Taylor AM (2004) Cardiac anatomy revisited. J Anat 205:159-177], one convention of the human anatomy is that all structures should be described in the setting of their anatomical position within the body and of the relationship of organs to each other. This convention has not always been strictly applied when describing the heart. In the past, the so-called Valentine approach has prevailed, that is, the convention of representing the heart isolated from the surrounding structures and balanced on its apex, with the atria above the ventricles. This approach generated confusion, particularly in the field of congenital heart disease, where the position of the heart and the location of the cardiac segments is variable. Therefore, as Prof. Anderson wrote: "students should be introduced to the anatomy of the heart as it lies within the body, as revealed with clinical tomographic images". To this purpose, the use of tomographic sections of isolated hearts, frequently used in textbooks of echocardiography, is only partially effective in prenatal ultrasound examination, where the views of the surrounding structures are much wider and the approaches to the fetal thorax more variable than in the postnatal setting.

We decided, therefore, to perform image sections of the whole fetal body to obtain tomographic views of the heart, thus showing the relationship between cardiac and extracardiac structures. As indicated in Chapter 21, tomographic sections of the fetal body were obtained in a limited number of fetuses under the gestational age of 20 weeks following informed consent of parents and in strict adherence with the Italian legislation on this subject. The idea to obtain these types of sections stemmed from almost 20 years of fundamental work by Alf Staudach [Staudach A (1989) Sectional fetal anatomy in ultrasound. Springer], with the advantages of computer technology in photographic and ultrasound images.

Rather than systematically describing congenital heart diseases in the fetus, the goal of this work is to provide a basic tool for understanding the normal and abnormal echocardiographic anatomy of the fetal heart. In this way, this textbook is complementary and not alternative to the more extensive publications on this subject.

The first section of this book describes the basic principles of diagnosis, illustrating assessment of the laterality of the fetal body, the visceroatrial arrangement, and the cardiac position. This order reproduces the logical sequence the examiner should follow when studying the fetal heart. In the second section, all echocardiographic projections in the fe-

tus are described, from those most familiar to obstetric sonographers, to those usually obtained only by pediatric cardiologists. Particular emphasis is given to imaging the short-axis sections of the fetal body, which has recently proven to be a powerful method for complete examination of the fetal heart. The chapters pertaining to echocardiographic projection are in logical sequence in two series: the transverse views of the fetal thorax, presented from the bottom to the top, and the sagittal and parasagittal views, presented from the right side to the left side. The sequence of these planes in some ways imitates the changes of the scanning plane that can be obtained by the examiner tilting or translating the probe manually with traditional probes or electronically with modern 3D ultrasound.

The third section consists of two chapters. The first describes the essential use of modern 3D/4D ultrasound techniques to image the fetal heart, including new volume manipulation, such as spatial and temporal image correlation (STIC), and rendering, such as glass-body, minimum-transparent, and inversion modes. The second chapter emphasizes the importance of autopsy in providing information regarding anomalies of the fetal heart and describes optimal techniques for dissection and photography of the autopsic specimens. This section emphasizes the crucial role of the echocardiographic projections described elsewhere in this book, which maintain their fundamental role in the comprehension of new advances in ultrasound and the future application of MRI/CT in postmortem studies of the fetal heart. These two different chapters are incorporated in this section because the approach to volume data sets of 3D/4D sonography and to blocks of pathological specimens shows many similarities. The information displayed by either technique depends on the level at which the examiner cuts the cardiac volume, no matter if a digital tool or a pathologist's blade is used. Moreover, some image-rendering techniques are comparable with the fine-art photographic technique available to the pathologist to display with outstanding clarity the subtle details of congenital heart disease in fetal heart specimens.

The problem of image orientation was thoroughly discussed with cardiac morphologists Andrew C. Cook, Gianni Botta, and Robert H. Anderson from the very beginning of the editorial phase of this book. The reader will notice that some anatomical images of the transverse sections of the thorax do not "at first sight" match the echocardiographic images. We decided to maintain the echocardiographic images in their original caudocranial orientation, a standard that is accepted in MR and CT imaging, rather than flip them horizontally, which would reduce the resolution of the digital clips on the accompanying DVD. We decided, instead, to display a compass with every image, thus illustrating its orientation.

As to the abnormal heart, rather than fully describe a single congenital heart disease in different projections, the book and DVD describe how a specific projection appears in the normal heart and in some abnormal conditions. Although we understand that this is not sufficient for a comprehensive assessment of a specific disease, we believe this type of presentation reproduces the usual approach of the examiner, who is initially unaware whether the case being examining is normal or abnormal. The more the examiner understands the normal anatomy, the easier it is to recognize what is wrong.

Ultrasound assessment of the heart, whether in fetal life or postnatally, is based on moving images, and books therefore have limited value in teaching echocardiography. With recent advances in ultrasound systems, storing multiple digital frames and clips with superb image quality has become a reality. These advances have brought innovative applications to the clinical field and can be utilized as powerful multimedia presentations for teaching purposes. The accompanying DVD is such a tool. This is a complex DVD that includes more than 300 clips of normal and abnormal views of the fetal heart frames of each topic, with 11–13, representing a single cardiac cycle, each of which is displayed in a loop. The DVD in many respects is not simply a copy of the book. The text window contains essential information of the case being presented, and – in the four-chamber section in particular – a short unit describes the essential features of each specific congenital heart disease. We believe this presentation will be a highly useful tool for all those interested in the echocardiographic study of the fetal heart.

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First of all, a special tribute to Prof. Robert H. Anderson, a giant in the field of morphology in congenital heart disease, whose teaching through the countless numbers of papers, books, lectures, and courses deeply impacts the practice of modern pediatric cardiology and cardiac surgery and, inevitably and unavoidably, the pages of this book.

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Florence, August 2008 E.M. Chiappa

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PART I

Basic Principles of Diagnosis

CHAPTER 1 General Guidelines

Introduction

Congenital heart diseases are the most common congenital malformations, affecting six to eight per 1,000 live births [1-3], and their prevalence in abortuses has been shown to be even higher [4, 5]. Moreover, heart diseases are the leading cause of death among infants with congenital anomalies, causing nearly 20% of neonatal deaths and up to 50% of infant deaths due to congenital anomalies. Recent studies have demonstrated a positive impact of prenatal diagnosis in morbility and mortality rates in specific groups of congenital heart disease [6-8]. Only 20% of congenital heart disease in the fetus occurs in high-risk pregnancies; therefore, routine screening of all pregnancies is necessary. Prenatal screening cannot be conferred solely to pediatric cardiologists because of the limited number of these specialists. The alternative, chosen in many developed countries, is to assess the heart in a simplified form – the four-chamber view – during routine "anomaly scan" at 18-20 weeks' gestation and to provide extra training for all obstetric sonographers. The anticipated potential of the four-chamber view to detect most of the severe cardiac anomalies has been disproved by discouraging false-negative rates of congenital heart disease in many screening programs utilizing this projection only [9-11]. Most forms of conotruncal anomalies, such as transposition of the great arteries, tetralogy of Fallot, truncus arteriosus, and double-outlet right ventricle, may appear completely normal in the four-chamber view. Some screening programs have demonstrated that the detection rate for congenital heart disease is significantly improved by an extended cardiac examination [12-16]. Therefore, there is a growing consensus that assessment of the ventriculoarterial connection should be included in the routine fetal anomaly scan [17, 18]. Detailed fetal echocardiography may identify most significant congenital heart diseases, but it is time consuming and requires special knowledge of the normal and abnormal cardiovascular system, which is not a requirement for every examiner. Most authors agree that to improve results in fetal echocardiography, efforts are needed to expand the skills of the operator involved with a clear and effective educational method. Positive results of training sonographers to recognize cardiac abnormality in the fetus have been proven [19, 20]. We believe the information provided in this work will be most useful to that end.

The background of this project comes from two main considerations. First, whether in fetal life or postnatally, echocardiographic diagnosis is based on moving images. Consequently, books have limited value in teaching echocardiography, and the use of videotapes is time consuming and relies on inconsistent image quality. With recent advances in ultrasound systems, storing multiple digital frames and clips with superb image quality has become a reality. These advances have brought innovative applications into the clinical field and can be utilized in powerful multimedia presentations for teaching purposes. Second, imaged sections of cardiac specimens are usually compared with their corresponding echocardiographic views in textbooks of echocardiography. These sections are mainly obtained from isolated hearts because they are technically easier and less time consuming to obtain. Nevertheless, imaged sections of the whole body are better tools by which to understand the relationship between cardiac and extracardiac structures. This understanding is particularly necessary in fetal echocardiography, where the number of visible structures around the heart is much greater and the approaches to the fetal thorax are more variable.

For this project, we created a digital presentation, included in the DVD, in which images from tomographic sections of the whole fetal body are combined with dynamic echocardiographic images of normal fetuses and of some of the most common congenital heart defects. All projections are accompanied by schemes and full text for better understanding. We believe this collection will be a major tool for all those interested in studying the fetal heart.